Reflex Sympathetic Dystrophy in A 10 Year Old Pediatric Patient: Case Report

10 Yaşındaki Pediatrik Hastada Refleks Sempatik Distrofi

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ABSTRACT Reflex sympathetic dystrophy is characterized by spontaneous pain, swelling, dysaethesia, and allodynia usually of a distal site of the affected extremity. Reflex sympathetic dystrophy is well recognised and most commonly seen in adults in the fifth to seventh decade, but there are several reports of the disorder in the pediatrics. Children presenting with painful limbs often have many investigations and the diagnosis may be considerably delayed. This results in prolonged distressing pain for the child, anxiety for parents, and difficulty with treatment. The majority of reports involve children in late childhood or early adolescence, with a mean age of onset of 12-13 years and a higher frequency in girls than boys. Here we present a 10 year old girl, show the characteristics of RSD.

Key Words: Reflex sympathetic dystrophy; pain; autonomic nerve block

ÖZET Refleks sempatik distrofi genellikle distal eksremitede spontan ağrı, şişme, dizestezi ve allodini ile karakterizedir. Refleks sempatik distrofi iyi bilinen ve sıklıkla yetişkinlerde 5 ile 7. dekadda gözüken, fakat çocuklarda az bildirilen bir bozukluktur. Ekstremite ağrıları olan çocuklarda bir çok incelemeler yapılmakta bu da tanıyı belirgin şekilde geciktirmektedir. Bu sürenin uzaması çocuğu ağrı yönünden sıkıntıya sokmakta, ailenin anksiyetesini arttırmakta ve tedavinin güç olmasına yol açmaktadır. Bildirilen raporların çoğunluğu geç çocukluk veya erken adolesan dönemde, ortalama başlangıç 12-13 yaş ve kızlarda erkeklere göre daha fazla oranda görülmektedir. Biz, burada refleks sempatik distrofinin karakteristik özelliklerini gösteren 10 yaşında bir kız çocuğunu sunduk.

Anahtar Kelimeler: Refleks sempatik distrofi; ağrı; atonomik sinir bloğu

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Reflex sympathetic dystrophy (RSD) is characterized by spontaneous pain, swelling, dysaethesia, and allodynia usually of a distal site of the affected extremity.[1,2] Other features relate to the autonomic nervous system and include cyanosis, mottling, sweating (which is less common in children), and reduction in temperature. The newer nomenclature, CRPS, includes a spectrum of entities (including RSD and causalgia) characterized by pain with severity or duration out of proportion to that expected.[3]

RSD is well recognised and most commonly seen in adults in the fifth to seventh decade, but there are several reports of the disorder in the pediatrics. In some cases, particularly if there is delay in recognition or treatment, more permanent or serious features may develop; these include muscle atrophy, demineralization of bone, and contractures of soft tissue around the affected joint. Children presenting with painful limbs often have many investigations and the

diagnosis may be considerably delayed. This results in prolonged distressing pain for the child, anxiety for parents, and difficulty with treatment.[4]

In the early stages of RSD, there are no specific laboratory tests available to help diagnose the condition, and the radiological and bone scanning findings characteristic in adults are often not seen in children.[5,6] The lack of diagnostic investigations, the relative lack of recognition of the condition, and the more variable clinical presentation present in children often result in a delay in diagnosis and treatment.

The incidence of RSD is not known in children, but it is more common in athletic girls.[7] It presents at an average age of 12 years in most cases, and rarely occurs before 10 years of age, generally the legs are affected. Early recognition of the condition is important in order to relieve pain adequately, avoid further deterioration through immobilization,[8] and offer psychological support. Idiopathic musculoskeletal pain syndromes have been described in patients as young as 3 years of age, though RSD has not been reported at this age. The majority of reports involve children in late childhood or early adolescence, with a mean age of onset of 12-13 years.[1,2,9-11] Here we present a 10 year old girl, show the characteristics of RSD.

CASE REPORT

An 10 year old girl presented to hospital with a 6 week history of increasing pain in her right arm. She had first seen her pediatric doctor with pain in her right arm but no history of trauma. She was diagnosed as RSD and refer to algology division. After admission to the algology division her history was taken, physical examination was performed and further evaluations had been made. At the time of admission her arm was cold and relatively red below the elbow with burning pain radiating from the elbow to hand. Also her hand's skin was flaking off and she was not able to make fist because of pain (Figure 1). Conventional analgesics were unhelpful. Her pain was evaluated and according to the visual analogue scale (VAS), as 6-7 out of 10.

On first examination at the pain clinic her arm was noted to be cold and her hand was relatively red and oedematous. She had allodynia (pain provoked by a stimulus that does not normally cause pain), dysaesthesia and paraesthesia up to her elbow. Extensor and flexor arm muscles were weak, but results from neurological examination were otherwise normal. The radiological examination revealed no pathologic findings. Clinical ortopedic and pediatric consultations were also revealed no abnormalities.

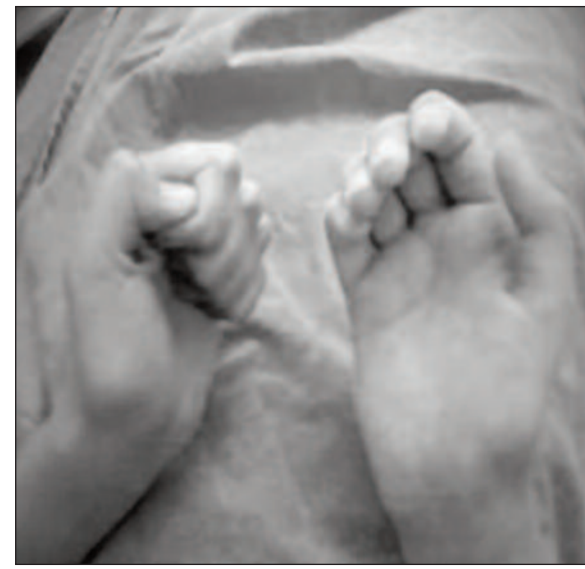

FIGURE 1: At the time of admission before procedure.

She was started 1 mg/kg tramadol gtt (Contramal gtt-Abdi Ibrahim/Turkey) and 30 mg/kg/day gabapentin (Neurontin- Pfizer/Turkey). Also, she was scheduled for right side stellate ganglion blockadge one week later.

Before the procedure her pain was evaluated and VAS was 4-5 out of 10. After the informed parental consent obtained, she was taken to procedure room. She recieved 0.5 mg i.v. midazolam before the procedure. The stellate ganglion block was performed under aseptic conditions with a 22 gauge blunt "B" bevel needle (Epimed - USA) as described in the technique section. Heart rate, pulse oximetry, and systolic and diastolic blood pressure were monitored. After the procedure she was taken to recovery room and observed for an hour. Five minutes after the procedure she was developed Horner syndrome but it resolved in couple of hours. 30 minutes later her VAS was 3-4 and she was discharged after one hour observation with no complication or any drug related side effects.

She was re-evaluated in the following week and VAS was 1-2 out of 10. Her arm was not cold.

Oedema on her hand was resolved and there was no radiating pain from elbow to hand. She did not have allodynia, dysaesthesia or paraesthesia up to her elbow. She stopped taking tramadol gtt.

In the following weeks, pain decreased gradually. During her last visit (for the following three months) she had no physical complaints. She was able to make fist normally with no pain in the hand and arm, and all the vascular changes and oedema had gone (Figure 2). Moreover, she was able move her arm without any pain. Dose of Gabapentin was gradually reduced.

TECHNİQUE

Each procedure was done by the same physician. The child was placed in the supine position and with the neck in slight extension. The physician performing the stellate ganglion blokadge was positioned at the child`s ipsilateral side of the reported pain. The seventh cervical vertebra was identified under fluoroscopic guidance. After sterile surgical conditions were obtained, a 22 gauge blunt "B" bevel needle was inserted and aimed towards the lateral side of the body of C7 (Figure 3). When bony contact was made and visualized on fluoroscopy, the needle tip lied deep to the anterior longitudinal ligament. Than the needle was withdrawn approximately 1 mm and, following a negative aspirate for blood and/or CSF, contrast was injected and deposited ventrolaterally to the body of C7 where the stellate ganglion is located and the spread of contrast media was towards the first thoracic sympathetic ganglion. Afterwards the child received 5ml of local anesthetic (5 mL of 1% lidocaine and 30 mg of triamsinolone [Kenacort-A, Bristol Myes Squibb] solution). One millilitre of local anesthetic was administered through the needle initially as a test dose, followed by a latency period.

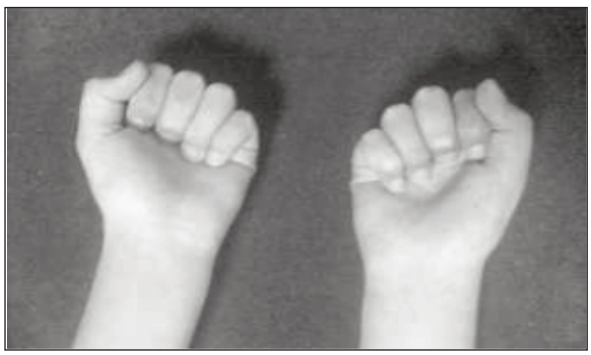

FIGURE 2: After procedure. She was able to make fist.

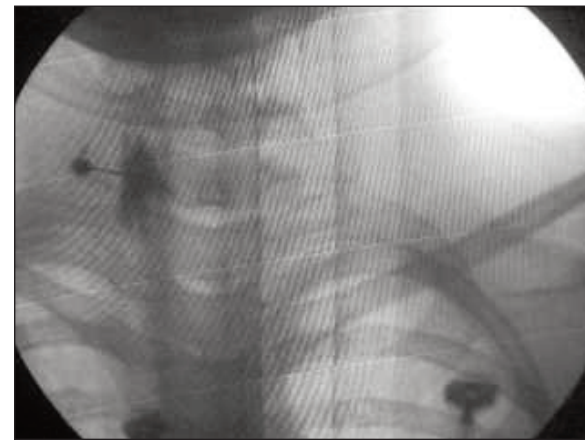

FIGURE 3: Stellate ganglion blokadge

DISCUSSION

RSD is a condition affecting both adults and children that results in significant morbidity. This condition is often challenging to manage, but can be treated very successfully with non-invasive but aggressive conservative management.[12] This is supported by the evidence provided in this study as well as other studies reported in the literature.[13-15]

The incidence of reflex sympathetic dystrophy in children is not known, but the disease is probably under diagnosed. Although the symptoms are the same in adults and children, the pattern of presentation is different. In adults it often occurs after a fracture or other trauma and follows a period of immobilisation. Incidence of RSD in girls is higher when compared with boys, as demonstrated in other studies.[13,16,17] Most cases occurred predominantly in the lower extremity compared with the upper extremity[16] and it is more common in athletic girls; and in most cases the legs are affected and there is no history of trauma.[9] In our case she was not dealing with sport, and her upper extremity was affected.

At presentation the autonomic signs may be subtle but are often present.[7] Investigations should include limb radiography, bone scanning, and thermography with thermal stress testing.[18] Patients should be referred early to a pediatric pain clinic where other diagnostic tests may be performed. All of these tests were performed in our case.

The main psychosocial indicator was the high prevalence of relatives in the same family with chronic pain conditions. There have been several psychosocial indicators related to RSD, and those identified were related to role models of pain and school stress. Our case was also seen by a pediatric psychiatrist to eliminate the other factors.

Many of the children had been treated with a period of immobilization; all of these children reported an increase in symptoms.[19] Considerable evidence supports the approach of using a multidisciplinary team rehabilitation therapy program to improve the outcome of children.[20]

Common therapies include physical therapy, nerve blocks, tricyclic antidepressant medication, opiate medication, anticonvulsant medication, and psychologic treatments.[21]

Many methods of managing this condition have been reported with varying success, including exercise therapy,[22] transcutaneous electrical nerve stimulation, sympathetic nerve blocks,[23] spinal cord stimulation,[24] and such medications as gabapentin,[25] calcitonin,[26] intratechal baclofen.[27] The majority of these studies include small numbers and actual controlled trials are lacking.

After we started analgesic and antiepileptic medication, we performed stellate ganglion blockage. We did not use a period of immobilization as a treatment bedsides we have observed a quick improvement at the movements of effected arm.

As a consequence, RSD is an important diagnosis in childhood and can cause considerable pain and morbidity. Although this diagnosis is rare, it should not be overlooked and the diagnosis should not be delayed. We think that besides the medical treatment sympathetic blocks should be considered in the management of RSD in children.

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